An unusual cause of acute lower gastrointestinal bleeding

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Abstract

A 45 year old male patient was referred to hospital after syncope during defaecation. Rapid peranal blood loss occurred shortly after admission. Gastroscopy and coloscopy performed as first line diagnostic measures failed to detect the source of haemorrhage. Ultrasound (US) revealed a hypoechoic and hypervascularized tumor mass in the right lower abdomen. A gastrointestinal stromal tumor (GIST) of the jejunum was diagnosed after laparatomy. Collectively US should be among the first line diagnostic procedures in younger patients presenting with lower gastrointestinal haemorrhage. (Acta gastroenterol. belg., **2006**, 69, **221-223**).

Introduction

Acute lower gastrointestinal bleeding (LGB) is defined as a bleeding of less then 3 days duration caused by a pathology distal to the ligament of Treitz and resulting in an instability of vital signs, anaemia and/or the need for blood transfusion (1). Haematochezia is the most common symptom of LGB however blood loss of more than 100 ml/h also occurs and may lead to rapid development of haemodynamic instability. The incidence of lower gastrointestinal bleeding is low compared to that of the upper gastrointestinal tract and has been estimated to affect about 25/100.000 per year with a strong increase with age (2,3). While endoscopy is the diagnostic measure of choice in chronic or occult LGB, patients with acute LGB are often not properly prepared and coloscopy is difficult, even for experienced endoscopists. Alternative procedures include ultrasound (US), computerized tomography (CT)-scan, angiography and 99mtechnetium red blood cell scintigraphy. If haemorrhage does not stop spontaneously or is not accessible through coloscopy, surgery may be the only option in the treatment of patients, especially if the source of the bleeding is located in the small intestine.

Case report

Here we report about a 45 year old male white patient with no known previous diseases who was admitted to hospital because of a first time syncope during defaecation. Shortly after admission haematochezia and peranal loss of fresh blood occurred. Laboratory analysis revealed a haemoglobin level of 9,9 g/dl which decreased to 6,0 g/dl within the following 7 hours. A source of haemorrhage could not be detected through gastroscopy or coloscopy. After stabilizing the patient by substitution of erythrocytes and volume he was transferred to the University Hospital for further treatment. On physical examination heart rate was 119 beats/min and blood pressure was 105/65 mm Hg. Chest and heart examination were normal. The skin was of pale color with signs of vasoconstriction. Haemoglobin was 6,2 mg/dl and ongoing peranal blood loss was present. Abdominal ultrasound revealed a hypoechoic solid mass measuring 4,6x4,4 cm in the right lower abdomen. Multiple color Doppler signals were noted in the tumor indicating a strong vascularisation (Fig. 1). Digital subtraction angiography (DSA) of the upper mesenteric artery confirmed the diagnosis of a hypervascularized tumor with signs of active bleeding and arteriovenous shunting (Fig. 2). Laparatomy was performed subsequently. A tumor was localized and resected along with 41 cm of jejunum (Fig. 3). Pathologic examination found it to be 5.7 cm in diameter and arising from the jejunal wall, eroding the luminal side of the intestineum. The histological appearance was composed of spindle cells arranged in fascicles with nuclear palissading. Immunhistologic examination revealed positivity for KIT (CD117, stem cell factor receptor)(Fig. 4), sm-actin and focally for CD34. Approximately 1% of tumor cells stained positive for the proliferation marker Mib1 (Ki67). Necrosis was not seen. The diagnosis of a gastrointestinal tumor (GIST) was made by the pathologist and based on localization, size and cellularity it was assessed to be of low-grade malignant potential.

The patient's post-operative course was without complication and no further bleedings occurred. A CT-scan performed 4 days after laparatomy showed a suspect hypodense focus in segment 8 of the liver however a subsequent relaparatomy with intensive examination of the organ failed to localize a metastasis. A CT of the thoracic organs as well as an abdominal CT 6 weeks after discharge of the patient were without pathologic findings.

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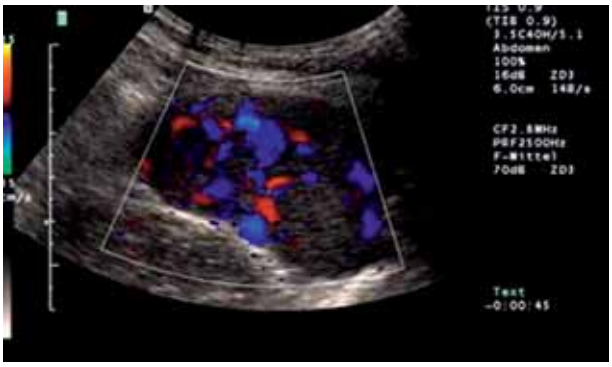

Fig. 1. — US of the tumor. Hypoechoic hypervascularized solid mass.

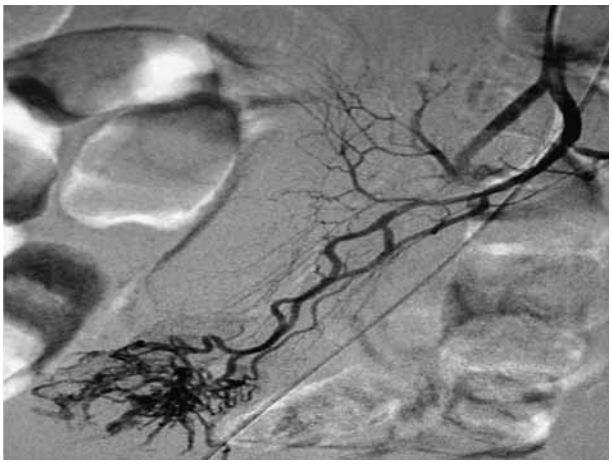

Fig. 2. — DSA of the mesenterial artery proving the hypervascularization of the tumor.

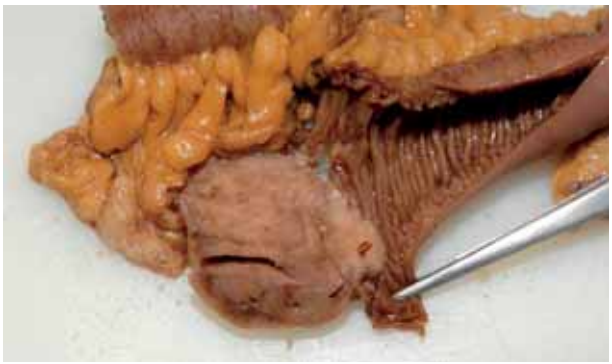

Fig. 3. — Resected jejunum with the extraluminal tumor arroding the intestinal wall.

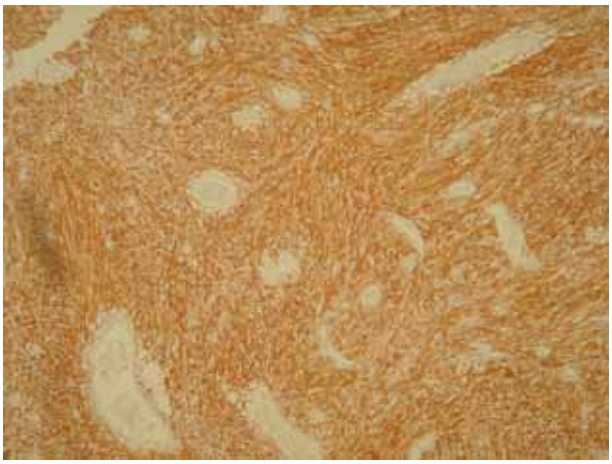

Fig. 4. — Immunihistologic positivity for CD117 (KIT)

Discussion

GIST represent < 1% of all primary gastrointestinal tumors (4). Although they may occur anywhere in the gastrointestinal tract, GIST arise mainly from the stomach (60%) and the small intestine (25%) (5). Extraintestinal localisations have been reported in rare cases and include the retroperitoneum and omentum (6). Gastrointestinal stromal tumors arise from interstitial cells of *Cajal*, autonomic nerve-related mesenchymal cells of the gastrointestinal tract (so called pacemaker cells) involved in the regulation of intestinal motility. They are generally defined as immunohistochemically KIT-positive mesenchymal tumors and it is the KIT protein, a transmembrane growth factor receptor for the stem cell factor (SCF), that is activated through mutation in GIST (7,8).

Numerous studies have been performed to define the dignity of GIST based on clinical and pathologic features. A localization in the stomach appears to have the best prognosis, whereas those in the small intestine and soft tissue of the abdomen have the worst. Malignant behavior of GIST is characterized by size, high mitotic activity, necrosis, infiltrative growth and marked nuclear atypia. Tumors with a maximal diameter of > 5 cm and a mitotic rate of five mitosess per 50 high-power fields (hpf) or more are usually regarded as malignant. Overall about 10-30% of GIST present a malignant behavior when initially diagnosed and have already metastasized or infiltrated adjacent tissues (9).

Of note the infiltration of the luminal side of the intestineum may be responsible for acute or occult blood loss. Therefore tumors of extraintestinal localisation are often not detected unless symptoms of compression occur. Several authors have reported cases of acute upper gastrointestinal bleeding caused by GIST arising from the stomach wall (10,11). In an 80 year old patient the application of diclofenac lead to a severe blood loss originating from a gastric ulcer eroding the submucosal tumor (12). The association between ulcerative colitis and GIST is discussed by Grieco et al. whereas Cueto et al. describe a patient in stable condition with persisting lower gastrointestional blood loss caused by a gastrointestinal stromal tumor (12,13). However, to our knowledge, a life-threatening acute LGB caused by GIST as in our patient has not been presented so far.

In an estimated 3-5% of patients with LGB the bleeding source is localized distally to the ligament of Treitz and proximal to the ileocolic valve. Meckel's diverticulum occurs in 1-3% of the population and is responsible for 30% of small-intestine bleedings in patients younger than 30 years (15). Aortoenteric fistula, vasculitis, diverticula or tumors (leiomyoma, carcinoid, lymphoma, adenocarcinoma) may, in rare cases, arise from the small-intestine and cause LGB. In many cases patients present with anaemia as the only clinical symptom. Endoscopy, contrast-enhanced CT, angiography and 99mtechnetium red blood cell scintigraphy are methods of choice to detect sources of intestinal haemorrhage. Lately capsule endoscopy has become a widely considered component of the diagnostic workup of obscure gastrointestinal bleeding (16). However in a situation in which rapid blood loss limits preparation of patients and the source of bleeding has to be determined quickly, endoscopy, CT, US, or explorative laparatomy may be the only diagnostic options.

Once the diagnosis GIST has been confirmed surgical resection is the therapy of choice. However even in patients with microscopically clean margins (R0) there is a high rate of tumor recurrence with a median time of 19 months between operation and tumor relapse. Radiation or conventional chemotherapy are barely options for treatment because of the relative resistance of GIST. Imatinib, a recently developed and orally applicable phenylaminopyrimidin-derivat, is able to block the specific ATP-binding site of tyrosin kinases such as KIT. Response rates are estimated to be between 37% and 51% (17).

In our patient gastroscopy and coloscopy failed to detect the source of a massive acute lower gastrointestinal bleeding in a 45 year old man. US was performed immediately after transfer of the patient to our hospital. In general tumors of 1 cm or larger are detectable by sonography, depending on localisation and abdominal gas. In the presented case it was US that revealed a hypervascularized tumor mass in the right lower abdomen leading subsequent angiography and laparatomy.

Collectively in younger patients with acute lower gastrointestinal haemorrhage, in which rare underlying causes must be especially considered, ultrasound should be among the first line diagnostic measures.

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